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Plasma cell mucositis

Dáire Shanahan MFDRCSI ¹ Deborah Shipley MRCP ² Konrad Staines FDSRCS¹

¹Department of Oral Medicine, University of Bristol Dental Hospital, Lower Maudlin St, Bristol BS1 2LY, United Kingdom.

²Department of Dermatology, Bristol Royal Infirmary, Upper Maudlin St, Bristol BS2 8HW, United Kingdom.

Correspondence to:

Dr. D. Shanahan
Department of Oral Medicine,
[University Of Bristol Dental Hospital](#),
Lower Maudlin Street,
Bristol, BS1 2LY.
UK.
Tel: +44 (0) 117 34 29641
Tel: +44 (79) 2847 2468
Fax: +44 (0) 117 34 24994
Email: daire.shanahan@UHbristol.nhs.uk

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A 62-year-old woman presented with a history of dry mouth, recurrent oral ulceration, periodic swelling to the upper right lip, dysphagia, and changes over the last two months to her voice. She sang in a choir and had problems vocalising both while singing and day to day voicing. She reported that she had lost some range when singing, and her voice was occasionally fatigable. Her inability to sing had greatly affected her quality of life. She had no skin, genital, gastrointestinal or ocular complaints, and did not smoke or drink alcohol. Her medical history included allergic rhinitis, occasional migraines, and gastro-oesophageal reflux disease (for which she took Omeprazole 20mg once a day).

Clinical examination revealed a diffuse erythematous and oedematous lesion on the soft palate, along with two ulcers in the left buccal sulcus (Figure 1). Various preliminary working diagnoses were given, including a lymphoproliferative disorder, sarcoidosis, granulomatosis with polyangiitis (Wegener granulomatosis), deep mycoses and syphilis.

Initial investigations comprised the following blood tests: FBC, haematinics, renal and liver profiles, C-reactive protein, immunoglobulins, serum & urine electrophoresis, antinuclear antibodies (ANA), anti-neutrophil cytoplasmic antibodies, serum angiotensin converting enzyme, complement, Hb1ac, syphilis serology, HIV, and a coeliac screen. Incisional biopsy and microbiology swabs were also performed; the biopsy showed inflammation with evidence of a dense plasma cell rich infiltrate. Polyclonality of the infiltrate was confirmed with kappa and lambda free light chain in situ hybridisation. A flexible nasendoscopy showed a spongiform appearance along the pharyngeal wall with significant oedema; however, the vocal cords themselves appeared unaffected. A diagnosis of plasma cell mucositis (PCM) was made.

Initial management was with systemic prednisolone 30mg. There was an improvement in symptoms and clinical appearance following the addition of Dapsone 50mg/100mg on alternative days. (Figure 2). Recurrent flare ups of symptoms resulted in prolonged systemic steroid use so mycophenolate mofetil 1gram twice a day was used in addition to the Dapsone. She has been relatively symptom free for the last year and remains under regular review.

Discussion

Plasma cell mucositis is a benign polyclonal plasma cell proliferative disorder of the mucous membranes with an unknown aetiology. The clinical appearances of PCM are varied, but typically it presents as a florid erythematous oral mucosa with cobblestone, nodular, papillomatous, granular or velvety surface changes. There is a slight male predominance of 1:2.1, and the average age of patients is 56.6 years. PCM is often associated with a synchronous or metachronous autoimmune or immunological dysfunction such as seronegative rheumatoid arthritis, Sjogren's syndrome, autoimmune hepatitis, polymyositis and diabetes mellitus.¹ There is one case report of invasive squamous cell carcinoma (SCC) developing from existing PCM of the lip.²

PCM can affect the oral cavity, as well as the nasal mucosa, nasopharynx, larynx, oropharynx, hypopharynx and esophagus. Patients can present with symptoms of oral pain, dysphonia, chronic cough, persistent hoarseness, dyspnoea, stridor, pharyngitis and dysphagia. Complications of PCM which require surgical intervention include tracheal strictures causing airway obstruction symptoms, along with debulking procedures of glottic and pharyngeal tissue causing dysphonia or dysphagia due to a mass effect.³

Clinicians must have a high index of suspicion for multiple myeloma and other plasma cell malignancies when considering a diagnosis of PCM. Evidence of monoclonal proliferation can be identified using serum protein electrophoresis (SPEP), immunofixation, and serum free light chains (sFLC), which together have a sensitivity of 97-98%. The principle histological features of PCM include epithelial hyperplasia and spongiosis, along with a dense polyclonal plasmacytic infiltrate in the superficial lamina propria. Failure to perform the above investigations can delay diagnosis of a plasma cell malignancy and have a significant impact on the clinical course of the disease and patients quality of life.

Management of PCM is challenging, and there is no consensus on treatment. The use of topical and systemic steroids is beneficial, but adverse side-effects limit prolonged use. Immunosuppressive agents have been used with varying success, including methotrexate, tacrolimus, dapsone, mycophenolate mofetil, cyclosporine, colchicine, azathioprine, infliximab, golimumab, and

adalimumab. Caution should be exercised when using potent immunosuppressive agents for this benign condition particularly where risk factors for malignancy exist.² Improvement in symptoms has been documented with low dose radiotherapy; however the follow up duration was short at twelve months. Surgical interventions are required in cases of subglottic stricturing or airway compromise, with two reported cases requiring tracheostomy.^{3,4}

It is important that PCM is recognised by head and neck practitioners, as its diagnosis is dependent on clinical and histopathological correlation.

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